

ESSENTIALS OF ECHOCARDIOGRAPHY #3

Heart Muscle Disease

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This unit covers the application of both M-mode and two-dimensional echocardiography in the investigation of heart muscle disease. A subsequent unit will discuss the application of echocardiography in congenital heart disease.

Left Ventricular Hypertrophy

Chronic elevation of myocardial stress due to pressure overload, as in hypertension or aortic stenosis, causes cardiac muscle to hypertrophy with a resulting increase in myocardial thickness. There is normally little or no change in overall cardiac size, so the wall thickening occurs at the expense of the cavity (**Fig. 1 and Fig. 2**). In the case of mitral regurgitation, on the other hand, ventricular volume increases with little or no change in maximum developed pressure; wall thickness does not alter significantly but total myocardial mass increases because of the ventricular enlargement. Except in the case of neonates, increase of mass does not alter the number of myocardial cells; the primary histological changes are intracellular, involving changes in the number and arrangement of the sarcomeres. In chronic cases widespread interstitial fibrosis occurs.

Echocardiography represents the method of choice for in vivo assessment of left ventricular hypertrophy. In particular, it is superior to both precordial palpation and ECG. These alternative approaches are relatively insensitive and subject to differences in observer skill and patient body habitus. A typical M-mode echocardiogram of left ventricular hypertrophy is shown in **Fig. 3**. Note the range marks are 1 cm, making the septum and the posterior wall of the left ventricle nearly 2 cm in thickness. A parasternal long axis (**Fig. 4**) and short axis (**Fig. 5**) from a patient with left ventricular hypertrophy show marked wall thickening in diastole. There is symmetrical thickening of both the septum and the free wall of the ventricle, and the cavity is small. The prominent papillary muscles are evident on the two-dimensional image. In systole, there may be virtual cavity obliteration due to the symmetric hypertrophy (**Fig. 6**).

In the majority of instances, left ventricular hypertrophy causes equal thickening of both the septum and the posterior wall. In perhaps 10% of cases, left ventricular hypertrophy may manifest on echocardiograms in an asymmetric form with the septum thicker than the posterior wall but without other stigmata of hypertrophic cardiomyopathy.

Echocardiography can be used to estimate left ventricular volume and mass. Since the two-dimensional images contain cross-sectional data, volume may be estimated from the short and long axes in systole or diastole. There are many mathematical approaches to the estimation of volume, the simplest based on the assumption that the geometric shape of the left ventricle roughly approximates a cylinder (for the base of the heart) placed on an ellipse (for the apical portions).

The formula for such a volume would be 5/6 the area of the short axis multiplied by the length of the ventricle (obtained from the long axis). Thus, the formula for estimation of volume is:

$$\text{Volume} = 5/6 \text{ Area} \times \text{Length}$$

Mass may also be approximated. The principle method is to estimate the total volume of the ventricle including the myocardium, and to subtract from this the volume of the cavity. This gives the volume of the myocardium, which is converted to mass by multiplying by the specific gravity of muscle (usually taken as 1.05). If the hypertrophy is symmetrical, and providing no regional wall motion abnormalities, these estimates show reasonable correlation with actual excised left ventricular weights measured at necropsy. Serial estimations of left ventricular mass can be used to monitor the efficacy of antihypertensive therapy or to determine the degree of myocardial recovery following aortic valve surgery. It should be noted, however, that such estimates require high quality two-dimensional images. In addition, these calculations are time consuming and not routinely performed in most laboratories.

Systolic function tends to remain normal in the presence of hypertrophy, but diastolic filling patterns are altered. Using M-mode studies with simultaneous apexcardiograms and phonocardiograms to delineate precisely the phases of the cardiac cycle, it has been shown that the protodiastolic isovolumic relaxation period is short and the ventricle fills slowly, peak filling rates of 5 cm.s^{-1} or less being not uncommon (normal $> 10 \text{ cm.s}^{-1}$). Diastolic hemodynamics are thus very similar to those of mitral stenosis, but the cause is the abnormal myocardium, whose relaxation is analogous to a stiff spring which exerts a powerful initial force but has a very limited range of motion. Now that Doppler echocardiography is available, such analyses of diastolic abnormalities are more readily derived from the diastolic flow patterns into the left ventricle through the mitral valve.

Severe pressure overload eventually leads to impairment of systolic function and finally to left ventricular failure. As systolic function deteriorates, the cavity enlarges, but the walls tend to remain thick. This is in contrast to the dilated cardiomyopathy of ischemic or idiopathic origin as discussed later.

Right Ventricular Hypertrophy

Right ventricular hypertrophy is commonly associated with any form of right ventricular outflow obstruction or pulmonary hypertension, which may in turn owe its origin to left-sided disease. The echocardiographic signs are thickening of the anterior right ventricular wall and the septum (**Fig. 7**). Cavity size is usually normal, or slightly enlarged. In many cases there is associated volume overload present due to tricuspid regurgitation, in the absence of this, septal motion is normal.

Unfortunately, M-mode echocardiography is insensitive for detecting right ventricular hypertrophy due to the lack of spatial information. Specificity is poor due to a number of factors, including variable orientation of the right ventricular walls to the ultrasound beam and the numerous other possible causes of septal thickening. Where some right ventricular hypertrophy is present, the papillary muscles attached to the right ventricular wall enlarge, and the path of the ultrasound beam may include these in the apparent septal thickness. Any condition affecting the right side of the heart tends to cause some cardiac rotation. The path of the ultrasound beam crosses a different region of the ventricle from normal, resulting in an apparent change in dimensions. Finally, there is the difficulty of an adequate 'gold standard' against which to judge echocardiographic findings.

Two-dimensional echocardiography overcomes many of these problems. **Fig. 8** shows a parasternal long axis in systole from a patient with right ventricular hypertrophy. The subcostal approach is a superior view for interrogating the right ventricle and overcomes the uncertainty of the beam pathway and may improve the specificity. Biventricular hypertrophy, difficult to analyze from ECG recordings, can be demonstrated dramatically by echocardiography (**Fig. 9**).

Left Ventricular Volume Overload

Two of the most common causes of left ventricular volume overload, mitral and aortic regurgitation, have been discussed in an earlier unit of this series. The basic feature of left ventricular volume overload is that the end-diastolic volume of the ventricle is increased showing a greater than normal cavity dimension at end-diastole.

Good myocardial function stimulates a powerful contraction. Through the Frank-Starling law the additional volume is ejected rapidly (though not necessarily into the aorta), and the end-diastolic dimension and volume remain normal. The relative reduction in cavity size, expressed as ejection fraction or shortening fraction, is therefore increased. Since ejection time is not prolonged (it may even be reduced if a lot of the blood is ejected into a low-pressure cavity), both the peak and mean values of Vcf, defined as reduction in chamber circumference divided by ejection time, are increased.

Much of this information can be deduced from simple inspection of an M-mode recording and confirmed by calculations derived from the cavity dimensions (**Fig. 10**). Wall thickness usually remains normal; this would be expected in the case of mitral regurgitation or ventricular septal defect, but is perhaps surprising in the case of aortic regurgitation where the increased volume must be ejected into a high-pressure cavity.

Most patients tolerate even quite severe left ventricular volume overload very well for many years, but eventually myocardial function becomes impaired. The only way in which a sufficiently large stroke volume can then be ejected is if the ventricle dilates still further. If this trend is unchecked, myocardial performance deteriorates rapidly, to the point where the risk associated with surgical correction of the original lesion becomes high. **Fig. 11** demonstrates a parasternal long axis from a patient with severe aortic and mitral regurgitations. The selected frame is in systole, showing marked left ventricular dilatation and very poor contraction.

Echocardiography contributes valuable, regular monitoring of ventricular function in such patients. Evidence of increasing systolic dimension should alert the physician to review carefully the other clinical signs and symptoms, particularly the patient's exercise tolerance. If the physician's review confirms the deterioration, surgery should be considered. Now that Doppler echocardiography is routinely available, the severity of chronic, or acute, left sided valvular regurgitation may also be followed. In this way surgical intervention can be considered before deleterious changes occur.

In most cases, however, a great deal of useful information may be deduced from the appearance of the left ventricle without Doppler. **Fig. 12** shows a systolic frame from a patient who had previously undergone an aortic valve replacement. The patient had done well for several years and presented acutely short of breath. The prosthetic valve had torn away from the annulus and was held by only a few sutures (**Fig. 13**). Note that the ventricular systolic diameter was increased (nearly 5 cm), indicating deterioration of ventricular performance.

Right Ventricular Volume Overload

Right ventricular volume overload is commonly caused by pulmonary or tricuspid regurgitation, or an atrial septal defect (not ventricular septal defect, as this presents the right ventricle with increased blood volume only after the onset of systole, therefore end-diastolic volume remains normal).

As in left ventricular volume overload, end-diastolic volume is increased and the force of contractions enhanced. This is reflected in the echocardiogram as increased end-diastolic dimension of the right ventricular cavity (an effect increased by rotation of the heart). The other striking echocardiographic finding is that the movement of the interventricular septum appears to reverse (**Fig. 14**). Instead of moving towards the left ventricle during systole, immediately after the ECG QRS complex, it begins to move anteriorly, then moves posteriorly again during diastole.

The mechanism of this reversed or 'paradoxical' septal motion was for a long time a subject of controversy. It is now fairly well established from two-dimensional studies that the major factor causing the apparent reversal is simply the overall motion of the left ventricle relative to the chest wall. Thus, in normal subjects the symmetrical inward contraction of the left ventricular walls is combined with an overall anterior motion during systole. This causes an apparent increase in the motion of the posterior wall and a correspondingly reduction in the motion of the septum. When right ventricular contraction is hyperdynamic, the anterior motion of the entire left ventricle becomes so great that there is actually a net anterior displacement of the septum relative to the ultrasound transducer. The shape and contraction pattern of the left ventricle itself remain normal.

Although most commonly associated with ventricular volume overload, paradoxical septal movement is found in several other conditions, (e.g., in the presence of a large pericardial effusion or following cardiac surgery). In such cases, this mechanism may be more complex: altered left ventricular function may play a part, as may the lack of restraint to cardiac motion if the pericardium is left opened after surgery.

Another case where a more complex situation exists is that of combined right ventricular pressure and volume overload, for example in severe pulmonary hypertension with secondary pulmonary and tricuspid regurgitation. On M-mode recordings the septal motion is strongly reversed, but has a much more 'square' appearance than in simple volume overload. Two-dimensional short axis views show the septum to be flattened so that the two chambers seem to exchange profiles: the right ventricle becomes circular while the left ventricle becomes more elliptical (**Fig. 15**).

Dilated Cardiomyopathy

Typical M-mode echocardiographic recordings from a patient with dilated cardiomyopathy are shown in **Fig. 16**. The small separation of the mitral valve leaflets suggests that there is low cardiac output. This appearance is known as the "fishmouth" configuration and is characteristic of such severe low flow states.

In some patients a 'hesitation' in the closure of the mitral valve leaflets at the onset of systole can also be seen (**Fig. 17**). Though the exact cause of this sign is uncertain, provided that the ECG P-R interval is normal, it is associated with significant elevation of left ventricular end-diastolic pressure. There is a lack of motion of the aortic root and reduced aortic cusp separation, which give further evidence of low cardiac output. The left atrium may be enlarged, due to elevation of ventricular filling pressure. As shown in **Fig. 18**, the left ventricular cavity is large with thin walls. Extreme dilatation of the left ventricle can stretch the mitral valve annulus and cause the valve to become incompetent.

In most cases of dilated cardiomyopathy there is a globular shape to the ventricle and wall motion is uniformly reduced. This can be visualized by two-dimensional echocardiography (**Fig. 19**). Note that there is little change in ventricular diameter from diastole to systole, a sign that overall left ventricular ejection fraction is quite low.

Hypertrophic Obstructive Cardiomyopathy

Echocardiography has been used extensively both as an aid to diagnosis and for research into the pathophysiology of hypertrophic cardiomyopathy (HOCM) otherwise known as idiopathic hypertrophic subaortic stenosis (IHSS). It is characterized by an abnormal arrangement of the myocardial cells, which instead of lying in parallel rows, form whorl-like patterns. It most commonly affects the interventricular septum, but may also involve the entire myocardium or occur in isolated areas undetectable except by detailed histopathologic examination.

In severe cases, the gross pathologic changes are striking (**Fig. 20**). The septum is massively hypertrophied, almost obliterating the left ventricular cavity and sometimes invading the right ventricle as well. The hypertrophied region often involves the left ventricular outflow tract, and causes obstruction to blood being ejected into the aorta. In a classic case, the septum is very thick, frequently over 2.0 cm, and usually moves poorly. The left ventricular cavity is small. The posterior left ventricular wall has normal thickness, and moves vigorously (**Fig. 21**). The left atrium is mildly enlarged as a result of elevated left ventricular filling pressure. The aortic valve cusps show an abnormal motion pattern, typically opening normally at the onset of ejection then fluttering to a semi-closed position (**Fig. 22**). This pattern is similar to that of a fixed sub-aortic obstruction (as described in an earlier unit of this series) but usually in hypertrophic obstructive cardiomyopathy the cusps tend to remain fully open for longer and the fluttering as they partially close is coarser and more irregular.

The motion of the mitral valve is also abnormal (**Fig. 23**). Because the ventricle is small, the anterior leaflet usually contacts the septum when it opens in early diastole. In addition, echoes that appear to arise from the mitral apparatus are seen to move up towards the septum during systole. The origin of these echoes, and the reason for the so-called systolic anterior motion (SAM) are controversial. It was initially thought that they arose from the mitral leaflets, but in many cases it appears from two-dimensional studies that they represent mitral chordae tendineae. One theory for the abnormal motion is that the high ejection velocity outflow tract causes sufficient pressures drop to suck upward the mitral valve through the Venturi effect.

In support of this argument is the observation that certain patients with hypertrophic obstructive cardiomyopathy do indeed have some mitral regurgitation. Another possibility is that the septal hypertrophy distorts the shape of the ventricle. **Fig. 24** shows a parasternal long axis from a patient with marked asymmetric septal hypertrophy and demonstrates that the left ventricular septal surface is concave in relationship to the cavity rather than convex as is normal individuals. **Fig. 25** shows the phenomenon of SAM where the chordae and leaflet tips approximate the septum in systole and result in left ventricular outflow tract obstruction. The thickened myocardium is often highly reflective of ultrasound in these patients and appears very bright on the two-dimensional echocardiogram.

None of the above findings is totally specific to hypertrophic obstructive cardiomyopathy. Thickening of the septum can be found in any condition that causes right ventricular hypertrophy, and left ventricular hypertrophy can be unevenly distributed in the myocardium to give disproportionate thickening of the septum. Infiltrative diseases can cause localized thickening of

the septum. It is also possible for the pattern of premature aortic valve closure to be indistinguishable from that of a fixed sub-aortic obstruction or even mitral regurgitation. The SAM pattern of mitral valve motion can be found in patients with mitral valve prolapse, where two-dimensional echocardiographic studies show the mitral chordae to have a whip-like action after the mitral valve closes at the onset of systole. Severe left ventricular hypertrophy can cause the mitral chordae to be displaced anteriorly during systole, though in this case they tend to follow the contour of the posterior endocardium.

In addition, there has been an excessive, and unwarranted, reliance on the fact that the ratio of the thickness of the septum to the posterior free left ventricular wall must exceed 1.3:1, as was published in the early echocardiographic literature. More recent studies have shown many different manifestations of this disorder, including severe concentric ventricular hypertrophy. Two-dimensional echocardiography has also identified another subset of patients where the hypertrophy is limited to the apical portions of the septum and free left ventricular walls. Careful examination of the apex is, therefore, requisite in all patients suspected of having obstructive cardiomyopathy. In these latter cases, SAM is frequently absent.

The diagnostic problems found in hypertrophic obstructive cardiomyopathy are a good example of one of the pitfalls inherent in any technique, namely of inferring that an echocardiographic abnormality reflects a particular pathological process. If all the features described above are present, it is most probable that the patient has hypertrophic cardiomyopathy; if only one or two are present, other possible causes should be investigated fully before this conclusion is drawn.

Even with these limitations, a careful echocardiographic examination is now the diagnostic procedure of choice for identifying patients with this entity. The constellation of findings of small left ventricular cavity size, asymmetric (localized or occasionally symmetric) left ventricular hypertrophy, highly reflective myocardium and SAM almost always means that this disorder is present. When SAM is absent, physiologic maneuvers or pharmacologic challenges may provoke this phenomenon and increase the diagnostic certainty. These include the administration of amyl nitrite or isoprenaline, and assuming an upright posture after a period of squatting. All are designed to reveal the presence of a latent dynamic outflow obstruction.

The extent of systolic mitral valve motion abnormality provides some indication of the severity of the obstruction. Two-dimensional studies often show that in mild cases the abnormal motion is confined to the mitral chordae, but with severe obstruction the extent of the abnormal motion increases and involves the mitral leaflets as well. In an attempt to quantify this observation, an "obstruction index", based upon the proportion of systole during which the mitral apparatus appears to be in contact with the septum, has been developed. However, although there is a moderately good correlation between this and the outflow gradient, the relationship is not close enough to be of much predictive value.

Echocardiography has virtually displaced all other diagnostic modalities for identification of hypertrophic obstructive cardiomyopathy. One of the additional contributions of echocardiography to our understanding of hypertrophic cardiomyopathy has been its ability to readily evaluate large numbers of patients and their relatives. Such studies show that the disease is genetically transmitted, by demonstrating its presence in asymptomatic relatives of patients known to have the disorder (the mode being autosomal dominant, with variable penetration). It has been suggested that the obstructive and non-obstructive forms of hypertrophic cardiomyopathy represent different diseases, but from an echocardiographic point of view the latter simply represents a milder form. The key finding is usually that of a thickened, relatively immobile septum with no other apparent cause.

Other Cardiomyopathies

The non-specific nature of the term “cardiomyopathy” allows a number of diseases directly or indirectly affecting myocardial function to be included under this heading. Many of these are rare, but the echocardiogram may be very useful in identifying patients with certain forms of cardiomyopathy. In general, echocardiography can detect any gross alterations they may cause in the structure or functioning of the heart, but such findings are likely to be non-specific.

Many cardiomyopathies are the result of infiltrative disorders that result in regional or global left ventricular thickening. One such disorder is sarcoidosis (**Fig. 26**), where the echocardiogram may reveal regional abnormalities. As a rule, however, most infiltrative cardiomyopathies affect all the myocardium. Amyloid infiltration of the myocardium usually presents a striking picture of marked left ventricular wall thickening (usually with normal cavity size), pericardial effusion and a very reflective myocardium (**Fig. 27**). While this appearance is non-specific, echocardiography provides a readily available method for identifying patients with infiltrative cardiomyopathy. Endomyocardial biopsy may be needed to provide more specific diagnostic information.

The fact that many infiltrative cardiomyopathies have similar echocardiographic manifestations is shown in **Fig. 28**. Here, a severely thickened and highly reflective myocardium is seen in an infant with Pompe's disease, a rare autosomally recessive inherited disorder of glycogen storage.

As is obvious from the echocardiographic appearance of these disorders, infiltrative cardiomyopathies are also referred to as "restrictive cardiomyopathies". The markedly thickened myocardium impairs its elasticity and results in significant difficulties of filling the ventricle in diastole but with more or less normal systolic contraction. As in the last figure that depicted a patient acutely dyspneic at birth, the echocardiogram frequently identifies genetic difficulty.

Some restrictive cardiomyopathies do not result from thickening of the muscle. Endocardial fibroelastosis (EFE) is manifest by marked endocardial thickening (**Fig. 29**) and normal (or thin) myocardium. This echocardiographic appearance is virtually diagnostic of this uncommon disorder.

Other, non-restrictive cardiomyopathies also occur. Entities such as idiopathic dilated cardiomyopathy result in global dilatation, wall thinning and hypocontractility that are echocardiographically the same as shown previously in **Fig. 19**. Severe ischemic cardiomyopathy may end in the same pattern. Inflammatory disease of the myocardium, or myocarditis, may be caused by a wide variety of bacterial and viral agents. Some forms are associated with specific cardiac abnormalities; for example, complete heart block in Chagas' disease and hydatid cysts in Echinococcus infection. The majority, however, do not cause specific echocardiographic abnormalities. The findings may mimic a dilated cardiomyopathy as a result of depressed myocardial function. Serial echocardiographic studies can be useful in monitoring the progress of the disease. Echocardiographic monitoring of left ventricular function is also of value in patients who are being treated with immunosuppressive or cytotoxic drugs.

Cardiac Tumors

Echocardiography is now the principle method for the diagnosis of cardiac tumors. As discussed in a previous unit in this series, myxomas are frequently intracavitary and may occur in any cardiac chamber. **Fig. 30** demonstrates a myxoma occurring on the tip of the anterior mitral valve leaflet of

an asymptomatic individual (who had an echocardiographic study as a part of an insurance physical).

There is, however, nothing specific about the echocardiographic appearance to identify the tumor. **Fig. 31** shows a left atrial tumor, attached to the posterior wall of the left atrium that on surgical resection was found to be an angiosarcoma. Since myxomas are most commonly attached to the interatrial septum, only its location on the posterior atrial wall indicated that it was unlikely to be a myxoma

Not all tumors are primary to the heart. **Fig. 32** shows a short axis of the aortic root with a massive tumor in the right atrium, protruding through the tricuspid valve orifice in diastole. This was found in a patient with diffusely metastatic melanoma who had a heart murmur noted on physical examination prior to chemotherapy. The patient had recently become intermittently dyspneic on exertion. The tumor was surgically removed to palliate symptoms

Metastatic tumors are most commonly intramyocardial and may be manifest as localized thickening within the myocardium. Such tumors can affect contractility or they may impinge upon the various cavities of the heart and obstruct flow.

However, primary tumors of the myocardium can occur. The most common tumor of children is a rhabdomyoma. **Fig. 33** shows a rhabdomyoma of the posterior left ventricular wall in a 14 year old. As with tumors elsewhere in the body, a tissue diagnosis is necessary for absolute certainty of the nature of the mass lesion.

Certain extracardiac tumors may affect the heart and be noted by echocardiography. Intrathoracic tumors of any origin can compress the cardiac chambers from outside the heart. Renal cell carcinoma may grow into the renal vein and thus into the inferior vena cava, obstructing venous inflow (**Fig. 34**).

Echocardiography is now the method of choice in identifying the presence and extent of both intracavitary and intramural tumors. Cardiac catheterization rarely adds diagnostic or more detailed anatomic information concerning these mass lesions. It is now widely accepted that surgical decisions can be based on the clinical situation and echocardiographic data.

Pericardial Effusion

One of the applications that first brought echocardiography to the general attention of clinicians was its unique ability to detect pericardial effusion. It remains the most sensitive technique for the detection of this disorder.

Around the heart there exists a potential cavity between the visceral and parietal layers of the serous pericardium, into which the heart is invaginated during its development. There is normally a little fluid in this cavity that acts as a lubricant for heart movement, but the two layers of the serous pericardium essentially remain in contact. However, if the amount of fluid increases as a result of exudate from the pericardium, hemorrhage or the inadvertent infusion of fluid, the two layers become more widely separated.

Fig. 35 shows a slow sweeping M-mode from a patient with a small posterior pericardial effusion. Behind the region of the left ventricle a strong echo arises from the fibrous pericardium, which is stationary and separated by a relatively anechoic space from the moving myocardium. At the level of the atrioventricular groove, the pericardial echo joins the posterior heart wall again. This is

because this region of the pericardial cavity, a cul-de-sac termed the oblique sinus, has little ability to expand due to its attachments to the pulmonary veins and so, although it forms a potential cavity, it rarely fills with fluid. This frequently is a helpful sign for detection of pericardial effusion, and the differential diagnosis of pericardial from pleural effusion, or other anechoic spaces behind the heart.

Small effusions are generally confined to the region behind the left ventricle when the patient is supine, and may appear to vanish when the patient sits up, as they drain to the apical region. For these positional reasons, the echocardiogram first detects small effusions posteriorly while the chest X-ray first manifests effusions anteriorly. The echo is performed with the patient recumbent while the lateral chest X-ray is performed with the patient upright.

Larger effusions, on the other hand, can be detected both anterior and posterior to the heart. In the case of very large effusions (**Fig. 36**), the heart may take on a swinging motion within the bag of fluid, resulting in a characteristic echocardiographic appearance. The ECG in such cases shows alternation of QRS complex amplitude, a finding virtually diagnostic of pericardial effusion. With such unusual motion of the entire heart, analysis of the relative motions of cardiac structures becomes impossible. Thus, the apparent presence of such echocardiographic features as mitral prolapse or reversed septal motion is liable to be artifactual, and must be reassessed after removal of the pericardial fluid.

M-mode studies permit some broad classification of the size of pericardial effusion, yet the extent and distribution of fluid surrounding the heart are better assessed by two-dimensional echocardiography, particularly in the case of postsurgical patients whose adhesions may confine an effusion to a localized region (sometimes causing it to remain undetected by a conventional M-mode examination), or when the effusion comprises blood following post-infarction myocardial rupture (“pseudoaneurysm”).

A complete two-dimensional echocardiogram from all transducer positions provides the most sensitive method available for detection of pericardial effusion. **Fig. 37** demonstrates a parasternal long axis in a patient with a small pericardial effusion. **Fig. 38** shows the heart suspended in a large pericardial effusion, seen both anteriorly and posteriorly.

The short axis view usually shows a symmetric distribution of the fluid around the circumference of the heart in effusions that are not loculated (**Fig. 39**). When effusions are smaller, or loculated, this symmetric distribution is no longer seen (**Fig. 40**).

Loculated effusions may be found anywhere in the pericardial space. **Fig. 41** shows an effusion confined to the oblique sinus of the pericardium, just posterior to the left atrium. **Fig. 42** demonstrates an effusion loculated behind the right atrium. A small bit of separation may be seen inferiorly (as in the subcostal views) with a normal amount of pericardial fluid.

Certain effusions may be seen to contain fibrinous material (**Fig. 43**). Such material is rarely encountered in effusions of viral, uremic or post-infarction origin. In the absence of a history of trauma (where blood may clot in the pericardium) or bacterial infection (where the purulent fluid may contain fibrin) such material is invariably found with effusions of malignant origin.

An echocardiogram is of inestimable value for patients with an enlarged cardiac silhouette on chest X-ray. It can readily differentiate between enlargement due to chamber dilatation, ventricular hypertrophy or effusion.

When a pericardiocentesis is contemplated, it is now prudent to perform such a procedure with echocardiographic guidance. The most practical method to approach such a procedure is to cover the transducer in a sterile sheath and direct the beam parallel to the needle. By visually interacting with the ultrasound image the pericardial space may be rapidly and safely entered, reducing risk of perforation of myocardium.

Large pleural effusions may mimic pericardial effusions. In such cases, it is necessary to identify the bright target of the pericardium. While this may be done by M-mode (**Fig. 44**), it is more easily accomplished by the two-dimensional approach because of the spatial target information available (**Fig. 45**).

Particular care is needed in the case of an effusion that has been present for some time, or one that has otherwise become organized. Its ultrasound characteristics then closely resemble those of the myocardium and so it will no longer appear to be echo-free. Moreover, the pericardium will no longer be stationary, but will move more or less parallel to the myocardium. Careful adjustment of amplifier gain and correct identification of all structures involved (the mitral chordae, epicardium, endocardium, and pericardium), plus confirmation of epicardial/pericardial apposition at the level of the atrioventricular groove are necessary to avoid misdiagnosis.

Fig. 46 demonstrates an area around the heart filled with multiple echo dense targets. In this case, the patient had pericardial edema due to metastatic lung cancer.

Chronic inflammation of the pericardium leading to calcification and adhesions to the myocardium can so restrict cardiac function that surgical removal of the pericardium becomes necessary. Unfortunately, direct detection of pericardial calcification by echocardiography is not normally possible. At first sight this is surprising, since calcium in valves is easily visualized. However, when such calcification is encountered between the transducer and the heart, little ultrasound penetrates to create images of adequate quality for interpretation.

One of the clinical features of constrictive pericarditis, namely paradoxical arterial pulse, can sometimes be demonstrated by echocardiography. As shown in **Fig. 47**, inspiratory increase of right ventricular stroke volume causes the septum to be pushed back, thus lowering left-sided stroke volume. While the echocardiogram may be helpful in the diagnosis of constrictive pericarditis, the findings may be very subtle. An otherwise normal echocardiographic appearance does not routinely exclude pericardial constriction, and further hemodynamic study may be necessary to establish this diagnosis.

In the setting of pericardial tamponade, the fluid may compress the right atrium and/or the right ventricle from within the pericardium, impeding filling of the right heart. While such right atrial or ventricular “collapse” may be found in patients in clinical tamponade it is not always present. It is best to make all observations possible from the echocardiogram, but to restrict the absolute diagnosis of tamponade to the physical examination, and/or other available hemodynamic data.

ISCHEMIC HEART DISEASE

The potential value of echocardiography in ischemic heart disease lies in two areas: assessment of left ventricular function and diagnosis of complications that arise as sequelae of infarction.

Unfortunately, many patients with ischemic heart disease are overweight or heavy smokers, and have large ‘barrel’ chests with hyperinflated lungs. These factors make complete echocardiographic visualization of the left ventricle difficult. Even a limited examination is at

times impossible in some patients. Such limitations must be borne in mind when assessing the potential clinical value of an echocardiographic study.

Left ventricular wall motion characteristics are altered by transmural myocardial infarction. The changes are usually regional and may be detected by echocardiography provided that the ultrasound beam traverses the affected area and that image quality is satisfactory. **Fig. 48** demonstrates an akinetic septum resulting from an anteroseptal infarction while **Fig. 49** shows akinesis of the posterior wall.

Two-dimensional echocardiography is usually superior for this purpose because its wider field of view provides the ability to locate and determine the extent of the infarcted myocardium. Cases of extreme systolic wall thinning and/or dyskinesia are readily apparent from inspection of sequential two-dimensional echocardiographic frames. **Fig. 50** shows extensive thinning and akinesis of the septum from a patient with a massive anteroseptal infarction.

Usually, such wall motion abnormalities are best evaluated by comparison of the diastolic and systolic images. **Fig. 51** shows paired short axis views from a patient with normal wall motion characteristics. Note the symmetry of the ventricle in both diastole and systole.

It must be kept in mind that the heart moves through the interrogating plane as it cycles through diastole and systole. This gross movement, even in normal individuals causes different regions of the myocardium to be interrogated in systole and in diastole and may lead to spurious interpretive errors (**Fig. 52**).

In addition, incorrect orientation of the short axis plane such that the ventricle appears elliptical rather than circular will cause any motion of the posteroseptal and lateral walls to appear exaggerated. Another common error in interpretation of wall motion abnormalities concerns the relative movements of the endocardium, epicardium and pericardium. In normal individuals, the movement of the epicardium and/or pericardium is much less than the endocardium. In patients with marginal image quality where only a portion of the endocardium is identified, the interpreter should not compare areas of endocardial movement to areas of movement where only the pericardium is visualized. This leads to the incorrect interpretation that asynergy is present in the poorly visualized segment.

If images of suitable quality are available, it is possible to analyze ventricular wall motion on a regional basis and to apply semiquantitative or quantitative descriptors to each segment of the myocardium. A number of simple and complex formats for performing such assessment have been proposed. **Fig. 53** illustrates the most simplistic format where the ventricle is divided into five segments: septal wall, anterior wall, posterior wall, inferior wall and apex. Using all the views possible, most wall segments can be located echocardiographically, albeit some with difficulty. Wall motion is then scored as normal, hypokinetic, akinetic or dyskinetic in each of the five segments.

The most reliable interpretation of these changes comes from assessment of the moving image rather than still-frames. An inferior area of hypokinesis is seen in the short axis of the left ventricle shown in **Fig. 54**.

Animal studies have indicated that wall motion abnormalities will occur almost instantaneously following coronary ligation. The utility of such an examination depends upon the clinical questions posed. For example, echocardiography may be used to assess the extent of damage in a patient with classic EKG and enzyme changes of a myocardial infarction. In patients without classical

changes, echocardiographic assessment may be helpful in influencing the clinical decision making process by detecting the presence or absence of wall motion changes.

Recent advances in computer technology have allowed for the digitization of a single cardiac cycle and for continuous loop replay to assist the interpreter in detecting wall motion abnormalities. This technology also allows for the placement of similar views side-by-side on the same screen before and after exercise. Since only one beat is required, it can usually be captured despite the patient's heavy breathing following peak exercise. Such features in instrumentation are referred to as "cine-loops".

Thus, echocardiography is now growing in use for the exercise evaluation of patients with suspected coronary artery disease using protocols roughly equivalent to those used for radionuclide evaluations. When image quality is adequate, echocardiography serves as a cheaper alternative that does not require the use of isotopes. Because no ionizing radiation is involved, patients may be evaluated more often depending upon the clinical situation involved. Multiple recent studies have indicated that this approach is as reliable as other methods and serves as a suitable alternative.

Complications Of Myocardial Infarction

Echocardiography can be of great assistance in the evaluation of a patient who, either in the immediate post-infarction period or later during recovery, develops complications.

Dressler's syndrome

The most common complication is Dressler's syndrome, a pericarditis typically occurring a few weeks after myocardial infarction. Echocardiography is valuable for detecting the presence of an effusion and for showing that any apparent increase in heart size is due to this and not to the development of a ventricular aneurysm. The appearance of the effusion is as previously discussed. **Fig. 55** shows a small pericardial effusion in a patient with new onset of pleuritic-type chest pain three weeks following myocardial infarction.

With a large effusion and impaired ventricular filling, cardiac tamponade may develop. The provisional diagnosis is usually made from the clinical signs, but rapid echocardiographic confirmation of a large effusion in the presence of tamponade physiology indicates the need for pericardiocentesis.

Ventricular Septal Defect

The finding of a loud pan-systolic murmur in a patient with an acute infarction is a serious complication, requiring prompt diagnosis and surgical treatment if the patient is to survive. The site of maximal murmur intensity may aid auscultatory diagnosis, but otherwise the differential diagnosis can be very difficult. This is especially the case if an old infarction or left bundle branch block make electrocardiographic determination of the infarction site impossible.

Post infarction ventricular septal defect most commonly occurs at the junction of the anterior and posterior portions of the septum, usually near the apex. It is frequently difficult to visualize on any single view. While visualization of the actual defect may be difficult, it may strongly be suspected by a severe wall-motion abnormality in the distal septum. Now that Doppler echocardiography is

available, the presence of a post infarction ventricular septal defect may be confirmed with certainty.

Papillary Muscle Rupture

As with post-infarction ventricular septal defect, discovery of a new systolic murmur following infarction suggests rupture of a papillary muscle leading to mitral regurgitation. Even the most experienced clinicians have difficulty in differentiating the clinical presence of ventricular septal defect from that of a ruptured papillary muscle. Echocardiography, in combination with Doppler methods, now serves as the principal means for the detection and differentiation of these two entities.

Rupture of a papillary muscle head also provides a striking echocardiographic picture. Flail chordae and the muscle head itself may be seen to be whirling around in the ventricle (as discussed in an earlier unit in this series) with the flail leaflet moving into the atrium in systole; within hours, the muscle head and chordae can become firmly tangled.

Left Ventricular Aneurysm and Thrombus

Following extensive infarction, the affected myocardium may become thin and fibrotic resulting in outward bulging of the ventricular wall during systole. Aneurysms may occasionally have mural thrombus within the aneurysmal segment (**Fig. 56**).

The presence of an aneurysm can sometimes be inferred from a slow M-mode sweep from the base of the heart toward the apex, in which the ventricular cavity size is seen to increase markedly toward the apex.

However, two-dimensional echocardiography has largely supplanted the M-mode approach. Aneurysms are most commonly found in the septal, apical, and lateral regions, and thus are best visualized using the parasternal or apical long-axis and four-chamber approaches. As well as the presence of a region where the wall is thin and dyskinetic, the best criterion for diagnosis of a true aneurysm is the presence of a “hinge”, where there is a transition from normal to akinetic or paradoxical wall motion. **Fig. 57** shows an apical four-chamber view with the heart normal in diameter at the base and a massive left ventricular aneurysm involving the distal two-thirds of the chamber.

Echoes suggestive of clot are frequently seen in the apical region, or attached to the septum, in patients with ischemic disease and poor left ventricular function. **Fig. 58** shows a layered mural thrombus at the apex while **Fig. 59** shows a multilobulated thrombus or thrombi at the apex. It has been shown that the multilobulated presentation of a mural thrombus, particularly one with rapidly moving intracavitary components, has a higher incidence of peripheral embolization. Data is still inconclusive as to the need for, or benefit of, anticoagulation in patients with mural thrombi in the setting of myocardial infarction.

Another presentation of mural thrombus is when the morphology of the thrombus is “web-like”. Such a thrombus is seen filling most of the left ventricular cavity in a patient with severe ischemic cardiomyopathy in **Fig. 60**. Such thrombi are, however, more commonly restricted to the area of the ventricular apex.

Mural thrombi are almost invariably associated with an underlying wall-motion abnormality. Occasionally, chest wall reverberations make adequate interrogation of the left ventricular apex quite difficult, and the diagnosis of apical mural thrombus is best left to experienced observers.

While the sensitivity and specificity of the echocardiographic diagnosis of ventricular thrombus are not known precisely, there is a growing abundance of data that this approach is the most clinically reliable method currently available.

Myocardial Rupture and Pseudoaneurysm

Rupture of the myocardium may also complicate myocardial infarction. Depending upon the size and location of the rupture, immediate death of the patient may ensue. Such patients obviously rarely survive long enough for the echocardiogram to be performed. There are no specific echocardiographic findings that indicate myocardial rupture. However, a large pericardial effusion in the immediate post-infarction period might suggest that rupture has taken place.

Some ruptures may occur over time and then seal locally within the pericardium. A left ventricular pseudoaneurysm is thought to originate following this series of events. Such pseudoaneurysms are differentiated from true aneurysms by their very narrow neck (**Fig. 61**) and they may frequently contain clot.

Pseudoaneurysms may occur on any left ventricular wall segment involved with a severe transmural myocardial infarction. When the rupture occurs near the junction of the septum with the posterior or anterior free walls, rupture may be into the right ventricle and also result in a ventricular septal defect.

Tissue Characterization and Myocardial Perfusion

The use of ultrasound for the detection of abnormal myocardium or its use in the evaluation of myocardial perfusion is still experimental. Early work has, however, shown some promise in this regard and some limited discussion is warranted.

In some medical applications of ultrasonic imaging, notably breast scanning, the use of gray-scale displays allows some differentiation of tissues on the basis of their ultrasound reflection, scattering or transmission characteristics. So far, little attempt has been made to apply such techniques to the heart. Echocardiographers have been primarily concerned with locating the positions of the major cardiac structures, and studying their motion patterns. As a result, most commercial echocardiographic machines are designed primarily to detect the strong specular reflections generated at interfaces between blood and muscle or connective tissues and to suppress weaker signals. Nevertheless, it is apparent that gross changes in tissue characteristics can be detected using some ultrasound machines. One reason differentiation between echoes of different intensities is difficult with black on white photographic process is that it can register a range of intensity of only 10:1. The original echo signals register a range of intensity of 1,000,000:1. An abundance of in vitro data using the raw ultrasound signals shows that the signal characteristics differ between normal and infarcted tissue. The application of these approaches in vivo is, however, limited.

Direct ultrasound imaging of the proximal coronary arteries is possible in some patients. While detection of stenotic lesions near the coronary origins is possible, the possibilities for diagnostic errors remains high except in the most experienced hands.

Early experimental work has shown that visualization of the distribution of coronary flow within the myocardium is possible. Such work is based upon the careful preparation of contrast materials

where small microbubbles of uniform size (from 2 to 4 microns in diameter) may be prepared. Such bubbles readily transit the coronary capillary bed and during their transit serve as strong reflectors of ultrasound. Such contrast "wash-out" approaches have shown to be safely performed in the experimental setting by direct injection of these materials into saphenous vein bypass grafts at surgery. The practical clinical implementation of this method for evaluating coronary perfusion remains to be determined.

Heart Muscle Disease (Figure Legends)

Fig. 1 Short-axis section of the left ventricle in a normal heart.

Fig. 2 Short-axis section of the left ventricle in a case of severe left ventricular hypertrophy.

Fig. 3 M-mode recording of a case of left ventricular hypertrophy showing symmetrical wall thickening and a small cavity. Note also the reduced rate of early diastolic filling resulting from increased wall stiffness.

Fig. 4 2-D parasternal long-axis view showing symmetric left ventricular hypertrophy and a large pericardial effusion.

Fig. 5 2-D parasternal short-axis view of the left ventricle during diastole. Left ventricular hypertrophy and a pericardial effusion are present.

Fig. 6 2-D parasternal short-axis view of the left ventricle during systole showing cavity obliteration.

Fig. 7 M-mode recording in a case of right ventricular hypertrophy. There is a small pericardial effusion that separates the echoes of the right ventricular wall from the chest wall echoes. Note also the thick interventricular septum.

Fig. 8 2-D parasternal left ventricular long-axis view showing right ventricular hypertrophy (arrowed).

Fig. 9 2-D subcostal four-chamber view showing right ventricular hypertrophy.

Fig. 10 M-mode recording showing left ventricular volume overload. The end-diastolic dimension of the ventricle is increased and contraction is vigorous.

Fig. 11 2-D parasternal left ventricular long-axis view showing left ventricular volume overload.

Fig. 12 2-D parasternal long-axis view showing both hypertrophy and volume overload of the left ventricle in a patient with an aortic prosthetic valve.

Fig. 13 2-D parasternal short-axis view at the aortic valve level. The valve has become dehiscant and is held only by a few suture threads.

Fig. 14 M-mode recording of right ventricular volume overload causing reversed (paradoxical) septal motion. The right ventricle is enlarged and the septum moves anteriorly at the onset of systole (arrowed).

Fig. 15 2-D parasternal short-axis view at the papillary muscle level showing right ventricular volume overload.

Fig. 16 M-mode recordings of the mitral valve (left) and left ventricle (right) in a patient with dilated cardiomyopathy.

Fig. 17 Normal and magnified M-mode recordings showing the pattern of mitral valve closure associated with elevated left ventricular end-diastolic pressure. There is an inflexion on the A-C slope and final closure occurs well after the ECG R-wave peak.

Fig. 18 Pathologic specimen in a case of dilated cardiomyopathy. Note the enlarged left ventricle with heavily trabeculated walls.

Fig. 19 2-D parasternal left ventricular long-axis view in diastole (left) and systole (right) in a patient with a dilated cardiomyopathy.

Fig. 20 Short-axis pathologic specimen of hypertrophic obstructive cardiomyopathy (HOCM).

Fig. 21 M-mode recording of hypertrophic obstructive cardiomyopathy.

Fig. 22 Normal and magnified M-mode recordings of the aortic valve in hypertrophic obstructive cardiomyopathy. In midsystole the leaflets partially close (arrowed).

Fig. 23 M-mode recording showing the systolic anterior motion (SAM) of the mitral valve (arrowed) associated with hypertrophic obstructive cardiomyopathy.

Fig. 24 2-D parasternal left ventricular long-axis view showing asymmetric septal hypertrophy.

Fig. 25 2-D parasternal left ventricular long-axis view showing hypertrophic obstructive cardiomyopathy.

Fig. 26 M-mode recording of a patient with cardiac sarcoidosis mimicking late-state hypertrophic cardiomyopathy. The patient is paced for complete heart block.

Fig. 27 2-D parasternal left ventricular long-axis view showing an amyloid cardiomyopathy with a posterior pericardial effusion.

Fig. 28 2-D parasternal short-axis view at the papillary muscle level in a patient with Pompe's Disease.

Fig. 29 2-D parasternal left ventricular long-axis view showing endomyocardial fibroelastosis (arrowed).

Fig. 30 2-D parasternal left ventricular long-axis view showing a myxoma of the anterior mitral valve leaflet (arrowed).

Fig. 31 2-D parasternal left ventricular long-axis view showing a sarcoma of the left atrium (arrowed).

Fig. 32 2-D parasternal short-axis view at the aortic valve level showing a right atrial melanoma (arrowed).

Fig. 33 2-D parasternal left ventricular long-axis view showing a rhabdomyoma within the posterior wall (arrowed).

Fig. 34 2-D subcostal view of the liver, inferior vena cava, and right atrium. There is a tumor seen within the inferior vena cava (arrowed).

Fig. 35 M-mode slow scan showing a small pericardial effusion. The fluid-filled region ends at the level of the atrioventricular groove (arrowed).

Fig. 36 M-mode recording of a large, tuberculous pericardial effusion. Oscillatory motion of the heart within the fluid causes variations of ECG R-wave amplitude.

Fig. 37 2-D parasternal left ventricular long-axis view showing a small pericardial effusion.

Fig. 38 2-D parasternal left ventricular long-axis view showing a large pericardial effusion.

Fig. 39 2-D parasternal short-axis view at the mitral valve level showing a moderate-to-large pericardial effusion.

Fig. 40 2-D parasternal short-axis view at the papillary muscle level showing a small pericardial effusion (arrowed).

Fig. 41 2-D parasternal left ventricular long-axis view showing a loculated pericardial effusion behind the left atrium (arrowed).

Fig. 42 2-D apical four-chamber view showing a loculated pericardial effusion behind the right atrium (arrowed).

Fig. 43 2-D apical four-chamber view showing a large pericardial effusion with fibrin strands (arrowed).

Fig. 44 M-mode recording from a patient with a small pericardial effusion and a large pleural effusion. The pericardium can be seen between the two effusions.

Fig. 45 2-D parasternal left ventricular long-axis view showing a small pericardial and large pleural effusion.

Fig. 46 2-D parasternal short-axis view at the level of the papillary muscles showing pericardial edema (arrowed) in a patient with metastatic lung disease.

Fig. 47 M-mode recording of a case of constrictive pericarditis. During inspiration the right ventricle enlarges and the left ventricle is forced to become smaller. On expiration the left ventricle expands. This mechanism underlies the clinical observation of pulsus paradoxus.

Fig. 48 M-mode recording of an anterior myocardial infarction. The septum is akinetic and the posterior wall moves vigorously. Note the high intensity of the septal echoes, suggestive of fibrosis.

Fig. 49 M-mode recording of a posterior myocardial infarction. It can be seen that the posterior ventricular wall is akinetic and that the septum moves vigorously. Compare with Fig.48.

Fig. 50 2-D parasternal left ventricular long-axis view showing thinning of the interventricular septum (arrowed) following myocardial infarction.

Fig. 51 2-D parasternal short-axis views at the papillary muscle level in diastole (left) and systole (right) showing normal wall motion.

Fig. 52 Diagram showing the effect of cardiac motion on 2-D short-axis images. A-diastole; B-diastolic image; C-systole; D-systolic image. The plane visualized in systole is nearer to the apex than in diastole.

Fig. 53 Diagram of left ventricular regional wall segments.

Fig. 54 2-D parasternal short-axis views at the papillary muscle level during diastole (left) and systole (right). Note the lack of movement in the inferior wall (arrowed).

Fig. 55 2-D parasternal left ventricular long-axis view of a small effusion (arrow) in a patient with Dressler's syndrome.

Fig. 56 Section of the apical region of the left ventricle showing an aneurysm partly filled with organized thrombus.

Fig. 57 2-D apical four-chamber view showing an aneurysm of the left ventricle.

Fig. 58 2-D apical four chamber view showing a left ventricular aneurysm with apical thrombus.

Fig. 59 2-D apical four-chamber view showing an apical pedunculated thrombus.

Fig. 60 2-D apical four-chamber view with inferior angulation showing a webbed thrombus (arrowed).

Fig. 61 2-D parasternal short-axis view of the papillary muscle level showing a large inferior pseudoaneurysm. The myocardial rupture has extending into the right ventricle resulting in a ventricular septal defect (arrowed).